Case Report

Primary bilateral plasmablastic lymphoma of the testis in a human immunodeficiency virus positive man

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Abstract

Human immunodeficiency virus (HIV)-related lymphomas are predominantly aggressive B-cells lymphomas. The most prevalent of the HIV-related lymphomas are diffuse large B-cell non-Hodgkin's lymphoma (NHL), which includes primary central nervous system lymphoma, and Burkitt lymphoma, whereas primary effusion lymphoma, plasmablastic lymphoma (PBL), and classic Hodgkin lymphoma are far less frequent. Of these, PBL is relatively uncommon and displays a distinct predilection for presentation in the oral cavity. In this manuscript, we report a primary testicular form of PBL in 44 year-old Border Security HIV positive patient who presented with bilateral testicular swelling of 1-year duration. On cytopathological and subsequent histopathological examination, the diagnosis of bilateral plasmablastic NHL was made. Extensive systemic work-up failed to reveal any disease outside the testes. Immune suppression rather than HIV itself is implicated in the pathogenesis of lymphomas. Herein, we report a case of PBL as AIDS-related malignancy presenting in testes and its correlation with CD4+ count.

Key words: CD4+ count, human immunodeficiency virus, plasmablastic lymphoma, testes

INTRODUCTION

Plasmablastic lymphoma (PBL) is an uncommon malignancy, described almost exclusively as an AIDS-related lymphoma. The recent WHO classification of lymphoid neoplasms addresses PBL under a separate entity, a neoplasm that shows diffuse proliferation of large neoplastic cells, most of which resemble B-immunoblasts and have immunophenotype of plasma cells.[1] PBL was originally described as a rare variant of diffuse large B-cell lymphoma (DLBCL) involving the oral cavity and occurring in the clinical setting of human immunodeficiency virus (HIV) and latent Epstein-Barr virus (EBV) infection.[2] Till date, only 2 cases of PBL of the testis in HIV-positive individuals have been documented in literature. We herein add a third case of primary bilateral PBL of the testis presenting as AIDS-related malignancy.

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CASE REPORT

A 44-year-old Border Security Forces personnel, presented with a history of a progressive painful bilateral scrotal swelling of 1-year duration. He had high-grade intermittent fever associated with weakness, loss of weight, and appetite. On examination, he had pallor. There was no history of any noticeable lumps in other parts of the body. General physical examination was unremarkable, with no palpable lymphadenopathy or hepatosplenomegaly except for visible bilateral testicular enlargement [Figure 1a]. There was no history of use of immunosuppressive medications. He was diagnosed as HIV + by ELISA method. In the initial appearance of testicular swelling, CD4+ count was

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103/mm^3 and improved with antiretroviral therapy to 250/mm^3. Ultrasound of the scrotum showed bilaterally enlarged testes of heterogenous echotexture and bilateral hydrocoele with enhancement on computerized tomography (CT) scan [Figure 1b]. Fine-needle aspiration cytology of both the testes was done and cytological smears showed atypical large lymphoid cells showing plasmacytoid and immunoblastic differentiation, having basophilic cell cytoplasm, eccentric nuclei with 1–2 prominent nucleoli [Figure 2a]. The possibility of large cell non-Hodgkin's lymphoma (NHL) with plasmablastic differentiation was suggested. Radiological evaluation with CT scan of chest and abdomen did not show any other focal lesion. Further evaluation, that is, bone marrow biopsy was done to exclude plasma cell dyscrasias, which was normal with no marrow infiltration. Serum and urine protein electrophoresis were normal. Based on progressive symptoms and increased size of the lesion, the patient underwent bilateral orchiectomy. Grossly the testes weighed 220 g and measured 8 cm × 5 cm × 3 cm and 6 cm × 4 cm × 2 cm with a large, white, fleshy, soft, slightly tan mass replacing almost the entire cut surface with no evident necrosis or hemorrhage [Figure 1c]. Microscopic examination showed diffuse sheets of highly atypical cells having large round nuclei, coarse chromatin, and 1 or 2 prominent nucleoli [Figure 2b]. There were foci of necrosis and mitotic activity was brisk. On immunohistochemistry, (IHC) tumor cells showed positivity for CD38 [Figure 2c], CD138 [Figure 2d], vimentin, and epithelial membrane antigen. Immunostains for CD45, CD20, ALK, PLAP, cytokeratin, and EBV were negative. Ki 67 index was 90%. The patient was diagnosed as having an extramedullary plasmablastic tumor most consistent with plasmablastic NHL. The patient was planned for chemotherapy, but because of his deteriorating functional status and low CD4+ counts, he died 1 month after diagnosis.

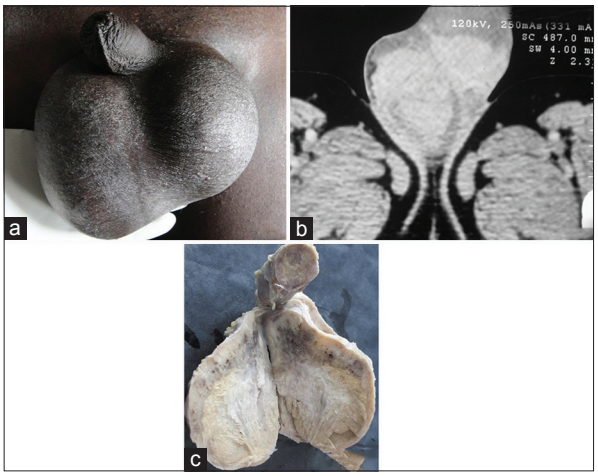

Figure 1: (a) Bilateral testicular enlargement (b) Computed tomography scan showing bilateral enlarged testes of heterogenous echotexture (c) Gross appearance of tumor, large grey white fleshy mass

DISCUSSION

Plasmablastic lymphoma, originally described in 1997 by Delecluse *et al.*, is an aggressive variant of diffuse large B-cell NHL seen predominantly in a setting of AIDS and nearly always in extranodal sites.[3] HIV infection significantly increases the risk for the development of lymphoma. Risk factors for the development of NHL in HIV include a low CD4+ T-cell count, high viral load, increased age, and male gender.[2] PBL accounts for 2.6% of all HIV-related NHL.[4] PBL usually develops in middle-aged adults, with the age at onset in one large series varying from 35 to 55 years.[4] It is characterized by immunoblastic morphology and plasma cell phenotype. In other words, plasmablasts are lymphoid cells that morphologically resemble B-cell immunoblasts but have acquired a plasma cell immunophenotype (i.e., loss of B-cell markers and surface immunoglobulin with the acquisition of plasma cell surface markers).[5,6] Main differential diagnoses include the plasmablastic variant of multiple myeloma and DLBCL with plasmacytic differentiation. Absence of monoclonal serum protein with no radiological evidence of lytic bony lesions favours the diagnosis of PBL. DLBCL shows positivity for CD45 and CD20. However, both of these markers are generally negative to weakly positive in PBL. Other morphological differential diagnoses which can be ruled out on the basis of IHC include Burkitt's lymphoma, lymphoblastic lymphoma, poorly differentiated carcinoma, and melanoma. To the best of our knowledge, only two cases of

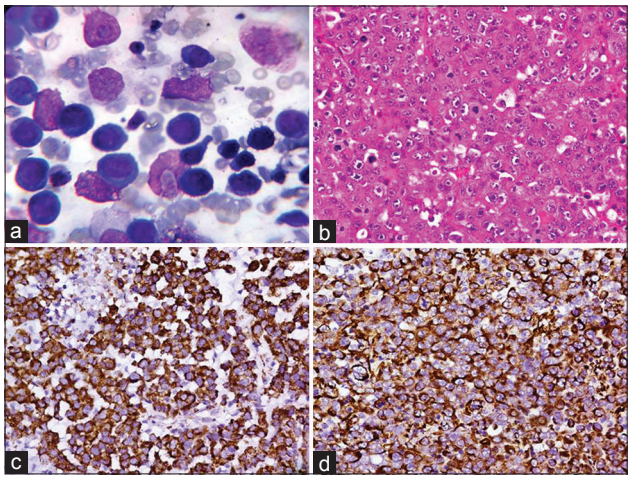

Figure 2: (a) Cytological smear showing atypical lymphoid cells with plasmablastic differentiation (MGG, ×400) (b) Microphotograph showing diffuse sheets of highly atypical cells having large round nuclei, coarse chromatin, and 1 or 2 prominent nucleoli (H and E, ×400) (c) Tumor cells showing immunopositivity for CD38 and (d) CD138 (IHC, ×400)

PBL of testis in HIV positive individuals have been documented in literature.[7,8] We are reporting the third case. Our patient was diagnosed to be HIV infected before his diagnosis of lymphoma and his CD4+ count was 103/mm^3 at the time of presentation. The unique features of our case were its bilateral testicular involvement without any appreciable lymphadenopathy. In the setting of HIV infection, these lymphomas tend to occur at a younger age, with higher histologic grade and apparent worse prognosis.

CONCLUSION

Human immunodeficiency virus infection-related lymphomas are uncommon. They are aggressive and mostly of B-cell immunophenotype. They tend to occur at a younger age and are associated with immunosuppression and low CD4+ count. The prognosis is worst. They respond poorly to treatment as compared to HIV negative lymphomas.

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